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The Department of Vermont Health Access Medical Policy

Subject: Huntington's Disease, Genetic Testing
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Revision 3:
Revision 2:
Revision 1:
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Description of Service or Procedure

Huntington disease (HD) is a brain disorder that affects a person's ability to think, talk, and move. It presents itself in three groups of symptom features: movement (chorea), psychiatric/behavioral and cognitive disorders. It is an inherited autosomal dominant condition meaning that each child of an affected parent, regardless of gender, has a 50% chance of inheriting the disease-causing gene. Some early clinical symptoms include dementia, mood swings, depression, lack of coordination, irritability, trouble driving, or learning new things. They may also have difficulty remembering a fact, or making a decision. As the disease progresses, concentrating and performing task becomes increasingly more difficult. The symptoms and severity of the disease vary between individuals as well as the age of onset.

The typical onset of HD symptoms is between ages 30-50. However, onset of symptoms has been seen in persons as young as 5 years or as old as 90 years. HD is associated with an expansion of a CAG tandem repeat. Analysis of the CAG repeat sequence genetic test is useful in three clinical situations: for confirmation of a suspected diagnosis of HD, for predictive testing in an asymptomatic individual known to be at-risk for carrying the gene, and for prenatal testing.

CAG REPEAT SIZE INTERPRETATION
26 and below Normal
27-35 Normal but potentially unstable
36-39 Abnormal with variable penetrance; unstable
40 and above Huntington Disease

Disclaimer

Coverage is limited to that outlined in Medicaid Rule that pertains to the beneficiary's aid category. Prior Authorization (PA) is only valid if the beneficiary is eligible for the applicable item or service on the date of service.

Medicaid Rule

7102.2 Prior Authorization Determination

7103 Medical Necessity

Medicaid Rules can be found at http://humanservices.vermont.gov/on-line-rules

Coverage Position

A genetic test for Huntington's disease may be covered for beneficiaries:

- When the genetic test for Huntington's disease is prescribed by a licensed medical provider, enrolled in the Vermont Medicaid program, operating within their scope of practice as described in their Vermont State Practice Act and, who is knowledgeable in the use of genetic test for Huntington's disease and provides medical care to the beneficiary AND
- When the clinical guidelines below are met.

Coverage Guidelines

Genetic testing for Huntington's disease may be covered for beneficiaries when:

- Diagnostic testing in an adult: To confirm or rule out the diagnosis. If symptoms strongly suggest the diagnosis of Huntington's disease but there is no known family history. Or
- Diagnostic testing in a child when symptomatic and suspected of having Huntington's disease based on family history and clinical phenotype. Testing of a child with the clinical phenotype in the absence of a family history should be done after ruling out other causes. It is not appropriate to presymptomatically test children (< 18 years of age) due to a parent diagnosed with adult-on-set Huntington's disease due to the inability to provide informed consent. It is appropriate to test children when symptomatic and suspected to have Huntington's disease (based on family history, clinical phenotype or both). Or
- Presymptomatic testing for a person at risk to provide reproductive and recurrence information.

And

- The results will directly impact the treatment plan of the beneficiary.
- Genetic counseling has been conducted.

References

Genetic Testing for Huntington's disease: Its relevance and implications (Revised). (2003). *United States Huntington's Disease Genetic Testing Group.* Retrieved January 29, 2016, from: http://hdsa.org/wp-content/uploads/2015/03/GeneticTesting-for-HD.pdf

Guidelines for genetic testing for Huntington's disease. *Hereditary Disease Foundation*. Retrieved January 29, 2016, from: http://dwb4.unl.edu/chem/chem869n/chem869nlinks/www.hdfoundation.org/testread/hdsatest.htm

Hayes, Inc. Hayes GTE Overview. *Huntington Disease for Huntington Disease (Various Manufacturers).* Landsdale, PA: Hayes, Inc.: April 2008.

Johnson, A.C. & Paulsen, J.S. (2014). Understanding behavior in Huntington's Disease. *Huntington's Desease Society of* America.Retrieved January 29, 2016, from: http://hdsa.org/wp-content/uploads/2015/03/Understanding-Behavior.pdf

Huntington disease. *Genetic Home Reference. U.S. National Library of Medicine.* Retrieved January 2, 2015, from: http://ghr.nlm.nih.gov/condition/huntington-disease

Huntington's disease. (2014). *Mayo Foundation for Medical Education and Research.* Retrieved January 2, 2015, from: http://www.mayoclinic.org/diseases-conditions/huntingtons-disease/basics/tests-diagnosis/con-20030685

Huntington Disease Mutation by PCR. *Indiana School of Medicine Department of Medical and Molecular Genetics. Division of Diagnostic Genomics*. Retrieved January 2, 2015, from: http://genetics.medicine.iu.edu/files/5414/0501/0324/HTT.pdf

Learning about Huntington's disease. (2011). *National Human Genome Research Institute. National Institutes of Health. U.S. Department of Health and Human Services*. Retrieved September 22, 2014, from: http://www.genome.gov/10001215

Myers, R. H. (2004). Huntington's disease genetics. *The Journal of the American Society for Experimental NeuroTherapeutics, 1.* Retrieved January 2, 2015, from: http://www.ncbi.nlm.nih.gov/pmc/articles/PMC534940/pdf/neurorx001000255.pdf

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